NIH RELAIS Document Delivery

NIH-10126337

NIH -- W1 AN62L

PAMELA GEHRON ROBEY
CSDB/NIDR/NIH Bldng 30 Rm 228
30 CONVENT DRIVE MSC 4320
BETHESDA, MD 20892

ATTN:		SUBMITTED:	2002-02-02 05:29:38
PHONE:	301-496-4563	PRINTED:	2002-02-04 10:35:59
FAX:	301-402-0824	REQUEST NO.:	NIH-10126337
E-MAIL:		SENT VIA:	LOAN DOC 5769542

NIH Fiche to Paper Journal

TITLE:	ANNALS OF PLASTIC SURGERY
PUBLISHER/PLACE:	Little, Brown And Company Boston Ma
VOLUME/ISSUE/PAGES:	1993 Jan;30(1):71-6 71-6
DATE:	1993
AUTHOR OF ARTICLE:	Tanaka Y; Tajima S; Maejima S; Umebayashi M
TITLE OF ARTICLE:	Craniofacial fibrous dysplasia showing marked invo
ISSN:	0148-7043
OTHER NOS/LETTERS:	Library reports holding volume or year 7805336 8333690
SOURCE:	PubMed
CALL NUMBER:	W1 AN62L
REQUESTER INFO:	AB424
DELIVERY:	E-mail: probey@DIR.NIDCR.NIH.GOV
REPLY:	Mail:

Craniofacial Fibrous Dysplasia Showing Marked Involution Postoperatively

Yoshio Tanaka, MD
Sadao Tajima, MD
Seiji Maejima, MD
Mie Umebayashi, MD

There have been few reports in the literature concerning the long-term prognosis of fibrous dysplasia. We reported here a boy with craniofacial fibrous dysplasia, which showed marked involution at the end of his puberty.

Tanaka Y, Tajima S, Maejima S, Umebayashi M: Craniofacial fibrous dysplasia showing marked involution postoperatively. Ann Plast Surg 1993;30:71–76

From the Department of Plastic and Reconstructive Surgery, Osaka Medical College Hospital, Osaka, Japan.

Received Sep 9, 1991, and in revised form Nov 25. Accepted for publication Nov 27, 1991.

Address correspondence to Dr Tanaka, Department of Plastic and Reconstructive Surgery, Osaka Medical College Hospital, 2-7 Daigaku-cho, Takatsuki, 569, Osaka, Japan.

Fibrous dysplasia is associated with facial deformity, exophthalmos, progressive visual loss, epiphora, and other such symptoms when it occurs in the craniofacial region. A conservative approach has been recommended because of its self-limiting growth, and surgical treatment is generally considered only for patients with a significant cosmetic deformity or a risk of optic nerve compression. However, there have also been reports that the radical excision of diseased bone with immediate craniofacial reconstruction is the best method of management, considering numerous examples of disease continuing beyond puberty and the risk of malignant transformation [1, 2]. We have treated seven patients with fibrous dysplasia affecting the craniofacial region. One of these patients, who showed pronounced shrinkage of the lesion at the end of puberty, is reported here.

Patient Report

An 11-year-old boy was first seen at our hospital in July 1984 with an 8-year history of progressive swelling of the right maxillo-orbital region. Protrusion of the right orbital region was first noticed at 3 years of age (1976), and epiphora developed due to involvement of the bony nasolacrimal duct. In January 1977 (at 4 years of age), biopsy was performed and the diagnosis of fibrous dysplasia was made at another university hospital. Because the disease was progressive and caused pronounced facial deformity, an attempt was made to remove the diseased bone in March 1977. However, most of the tumor remained untouched because of heavy bleeding, and radiotherapy was performed instead (2,400 rad).

Physical examination showed a large mass in the right maxillo-orbital region causing pronounced facial deformity (Fig 1). The right eye was displaced laterally and superiorly; there was proptosis of 6 mm, and the distance between the inner canthi was about 50 mm. The visual acuity was 0.06 on the right and 1.2 on the left. A posterior capsular cataract was seen on the right. The critical fusion frequency value was 30 Hz on the right side and 48 Hz on the left side. Examination of the fundi revealed only slight pallor of the optic disk, and the poor right visual acuity was diagnosed as being due to anisometropic amblyopia. Intraoral examination showed excessive bony growth of the right maxillary alveolar process and the hard palate, with the soft palate also being hardened.

Laboratory test results were normal except for alkaline phosphatase, which was elevated to 190 mU/ml (normal, 30–85 mU/ml). Plain radiographs showed an extensive osteosclerotic lesion on the right side of the face (Fig 2). A computed tomographic (CT) scan showed extensive fibrous dysplasia of the right maxilla, the medial orbital wall, and the sphenoid and ethmoid sinuses, with the right eyeball being displaced laterally and anteriorly (Fig 3A, B). Bone

A B

Fig 1. (A, B) Preoperative appearance showing massive protrusion of right maxilla and orbital region.

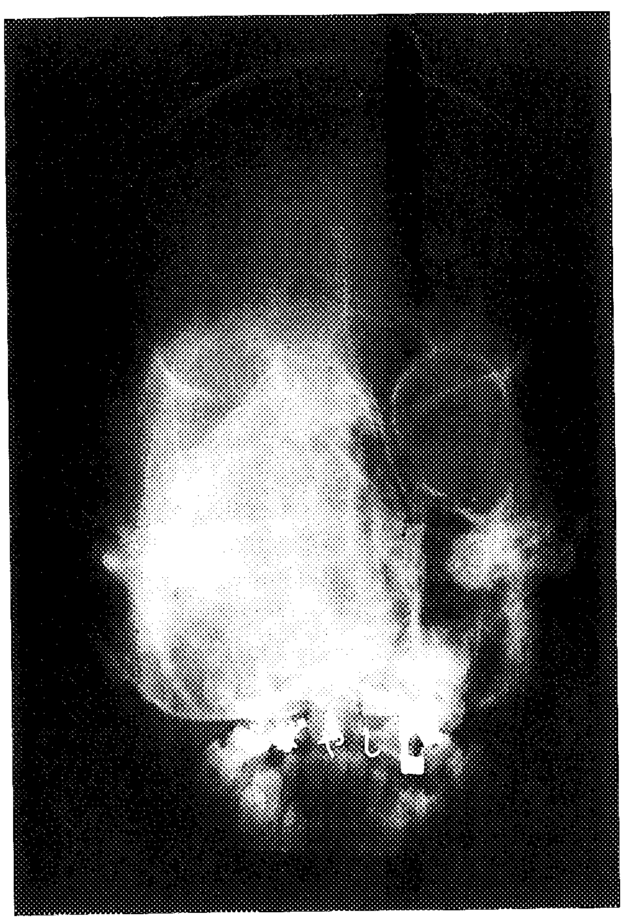


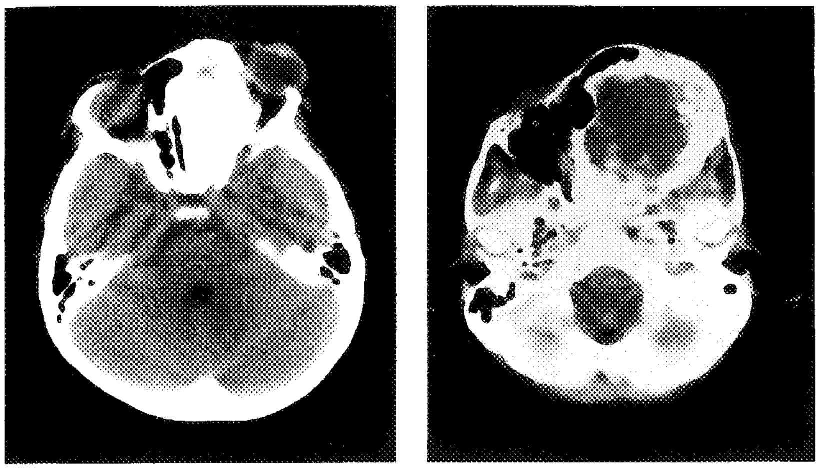
Fig 2. Plain radiograph revealing typical diffuse sclerosis of the facial bones.

scanning with ^{99m}Tc-methylene diphosphonate showed increased uptake around the maxilla (Fig 4).

Surgical Procedures

The first operation was performed in July 1984. The approach was made by excising the previous surgical scar along the lateral nasal area and through a lower eyelid incision. In the oral cavity, a full-thickness mucoperiosteal incision was also made in the right maxillary vestibule. Fibrous dysplasia was excised from the maxillary antrum, the frontal process of maxilla, the orbital floor, the medial orbital wall, and the hard palate. The tumor extending into the soft palate was also excised as completely as possible. The dysplastic bone had a consistency resembling soft sherbet and bled easily. The right nasolacrimal duct was freed from the surrounding dysplastic bone and placed into the maxillary antrum with its distal end opened.

The infraorbital rim was reconstructed using a graft fashioned from the excised dysplastic bone, and the medial canthal ligament was placed in its correct position by transnasal canthopexy (Fig 5A, B). Histological examination of the excised



A B

Fig 3. (A) Computed tomographic (CT) scan showing replacement of the right ethmoid cells by bony tumors. The right eyeball is displaced laterally and superiorly. (B) CT scan at the level of the maxilla showing massive bony proliferation with a central radiolucency, suggesting a fluid collection. Note the severe deviation of the nasal septum.

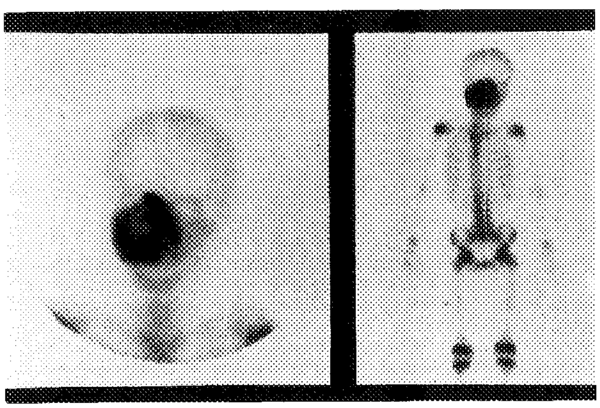


Fig 4. Bone scan with ^{99m}Tc-MDP; increased uptake is noted in the right facial region.

bone confirmed the diagnosis of fibrous dysplasia (Fig 6).

After the first operation, his cosmetic appearance was markedly improved and the problem of lacrimation was also relieved. However, because the right eye became somewhat enophthalmic with time (Fig 7A, B), further procedures to correct this enophthalmos, using iliac bone and costal cartilage grafts, were performed in July 1985 and August 1987, respectively. At the time of the second operation, we observed considerable resorption of the reimplanted dysplastic bone, and histological examination of this bone did not reveal any active fibrous dysplasia.

Postoperative Course

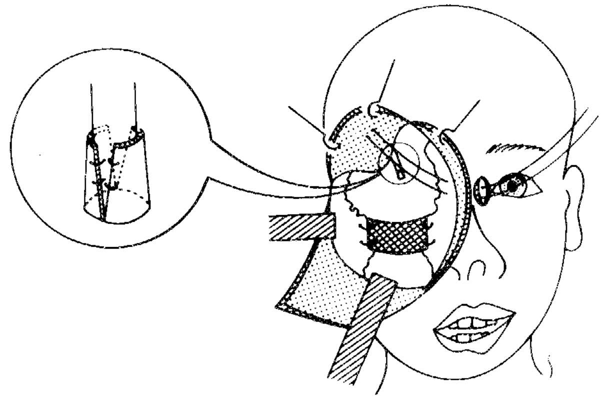
In November 1987, 3 months after the last operation, the boy showed slight shrinkage of the protrusion of the maxillary alveolar process where dysplastic bone still remained. By August 1990 (at 17 years of age; Fig 7C, D), the soft palate had become softer and the protrusion of the maxillary alveolar process had also disappeared (Fig 7E).

Since the first operation in 1984, the postoperative course was also followed with CT scanning. The ethmoid sinus became air-filled, and the bony lesions in the orbit also disappeared with time, resulting in enophthalmos (Fig 8). Although bone scanning with ^{99m}Tc-MDP still showed increased uptake around the maxillary region, its extent was reduced (Fig 9). These findings show that involution of the fibrous dysplasia is still in progress.

Discussion

The cause of fibrous dysplasia is not known and several theories have been proposed. Lichtenstein and Jaffe [3] suggested that "it is derived from the abnormal development of undifferentiated osteoplastic mesenchymal tissue," and this is considered to be the most reasonable hypothesis at present. The average age of onset is 10 years, and it is generally said that the progression comes to a

A



B

Fig 5. (A) Contouring of the maxilla and orbital decompression were performed while excising the diseased bone. (B) Schematic representation of the reconstruction; the nasolacrimal duct was everted at its distal end and replaced into the maxillary antrum.

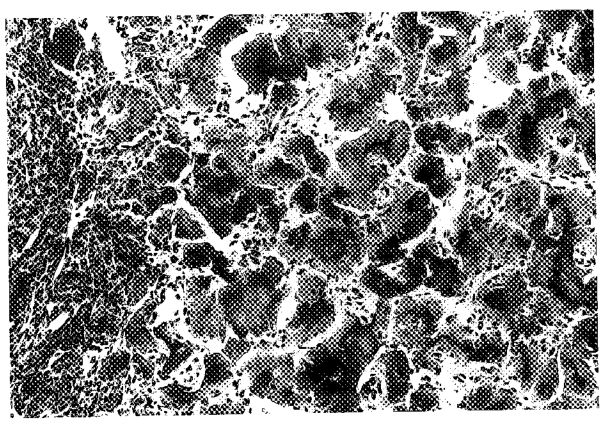


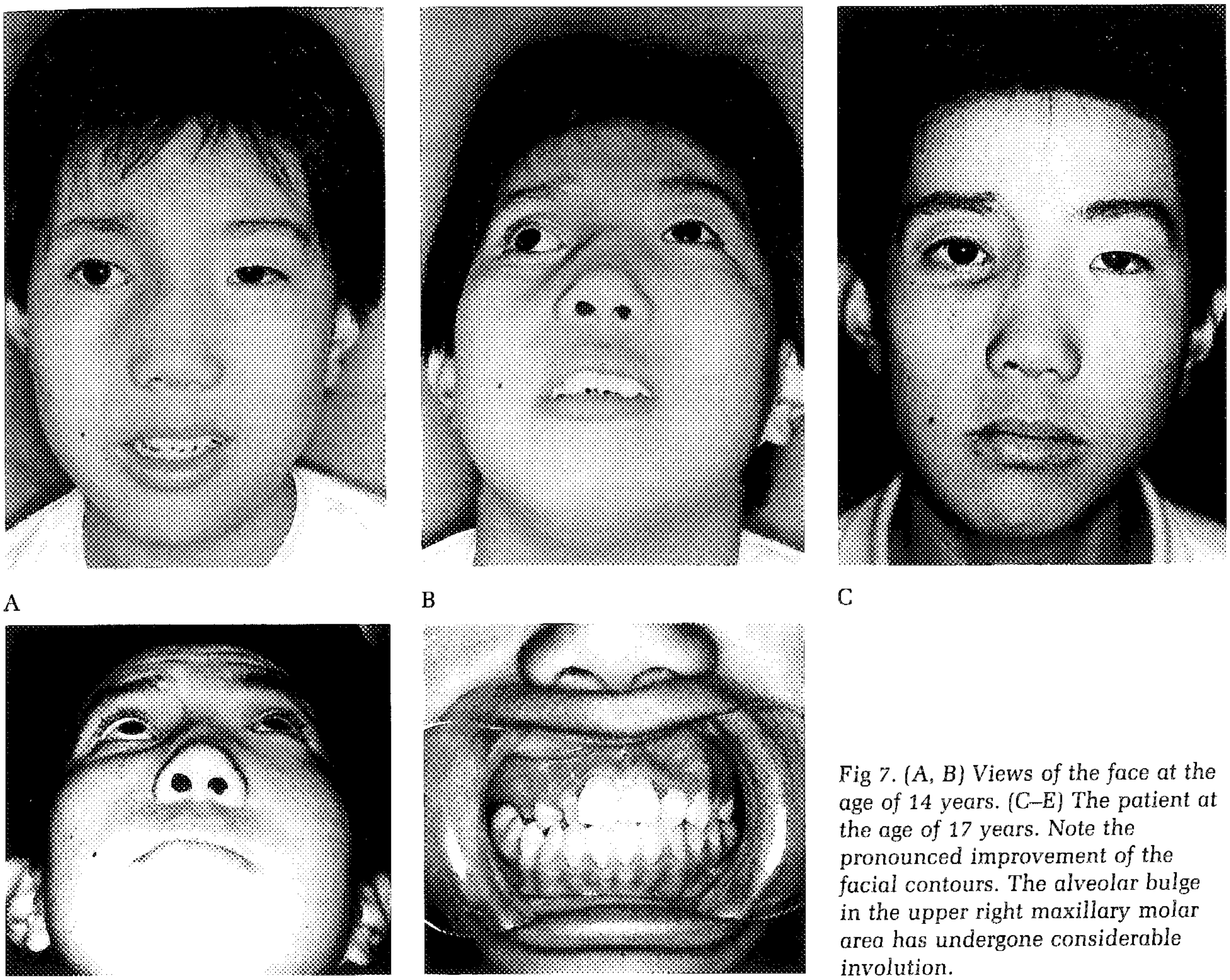
Fig 6. Fibrous dysplasia with proliferating fibroblastic tissue; the spherical masses of osseous tissue are surrounded by the proliferating spindle cells (hematoxylin and eosin, original magnification × 400).

relative standstill at puberty. However, there are also examples of the disease progressing beyond that period, especially when it arises in infancy or is polyostotic. The present patient had polyostotic disease that was first noticed at 3 years of age, so the disease was expected to be progressive for a long period. Nevertheless, postoperatively, the lesions showed gradual shrinkage and enophthalmos progressed unexpectedly.

There have been few reports in the literature concerning the long-term prognosis of fibrous dysplasia. Kanthak and associates [4] reported only one patient who showed shrinkage of the lesions 3 years after radiotherapy. Our patient also underwent radiotherapy at the age of 4 years at another hospital. Involution only became obvious, though, 10 years later, and was especially pronounced in the ethmoidal and orbital regions where irradiation had not been performed.

The most recent facial bone scan with ^{99m}Tc-MDP showed reduced activity in the right orbital and ethmoidal regions, whereas the maxilla, which received irradiation, still showed increased activity. Therefore, it appears that radiotherapy was not effective in producing involution of the lesions in this patient. The exact cause of involution is unknown, but the decompression of the enlarged antrum into the nasal cavity by excising dysplastic bone might have been a contributing factor.

In the surgical treatment, total excision of the diseased bone is ideal. However, excision may have to be limited, depending on the site and extent of the lesions. Because the patients are young and the lesion is a "dysplasia," pronounced postoperative deformity or functional defects should not be caused by surgery. In this patient, the diseased bone was excised within the scope of feasible reconstruction, and efforts were made to correct deformities while conserving function.



A B C D E

Fig 7. (A, B) Views of the face at the age of 14 years. (C–E) The patient at the age of 17 years. Note the pronounced improvement of the facial contours. The alveolar bulge in the upper right maxillary molar area has undergone considerable involution.

In reconstruction of the defects after surgical resection, cranial, iliac, or rib grafts have usually been used, but a method of remodeling diseased bone as a graft has also been reported [5]. In the present patient, the resected diseased bone was remodeled and used for reconstruction of the lower orbital rim. The reimplanted diseased bone did not grow again, and pronounced absorption was observed at the time of the following operation.

Finally, the malignant transformation of fibrous dysplasia to fibrosarcoma, osteosarcoma, or chondrosarcoma is very rare (less than 1%), but the frequency of postradiation sarcoma is high after radiotherapy. Edgerton and colleagues [6] reported that "the incidence of sarcoma is increased 400 times above the spontaneous rate in patients who have irradiation. Radiotherapy should never be used to treat fibrous dysplasia." However, in the present patient, it has already been performed, so careful follow-up is mandatory.

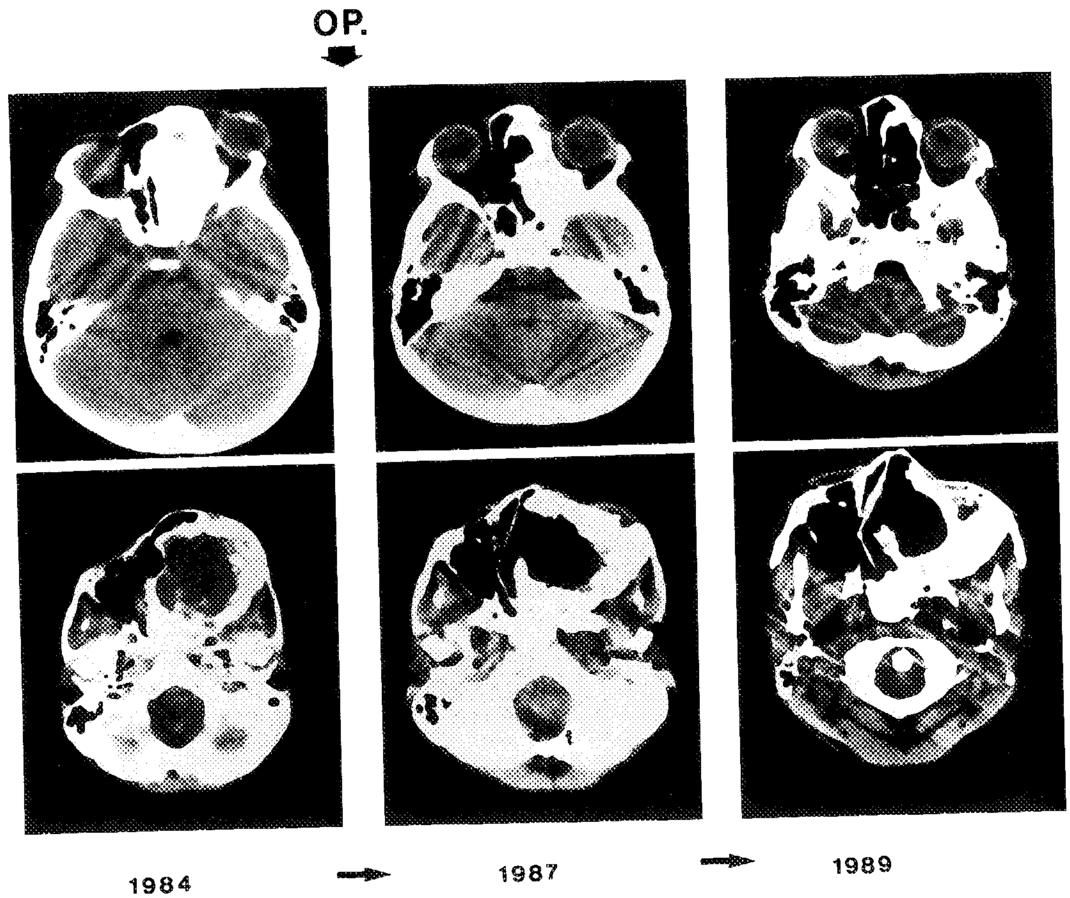


Fig 8. Computed tomographic scan at the level of the orbit and the maxilla. Note the involution of dysplasia, especially in the ethmoid cells and the orbit.

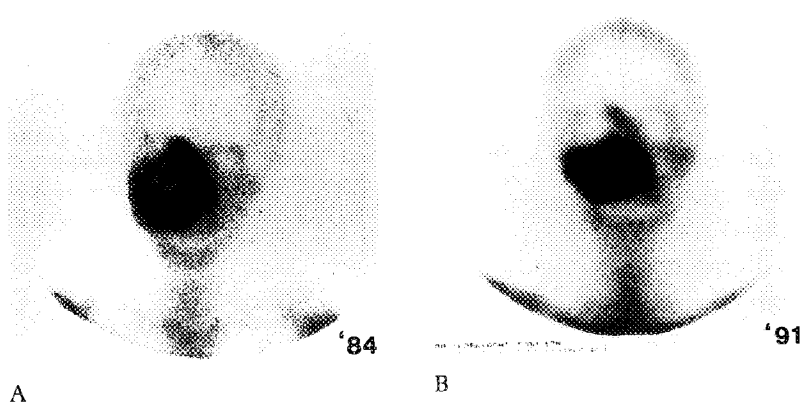


Fig 9. Bone scans with ^{99m}Tc-MDP. (A) Bone scan at the age of 11 years. (B) Another scan 6 years later at the age of 17 years.

References

1 Jackson IT, Hide TAH, Gomuwka PK, et al: Treatment of cranio-orbital fibrous dysplasia. J Maxillofac Surg 10:138–141, 1982
2 Moore AT, Buncic JR, Munro IR: Fibrous dysplasia of the orbit in childhood: clinical features and management. Ophthalmology 92:12–20, 1985
3 Lichtenstein L, Jaffe HL: Fibrous dysplasia of bone. Arch Pathol 33:777–816, 1942
4 Kanthak F, Hamm WG, Yarn CP Jr: Fibrous dysplasia of the facial bones. Plast Reconstr Surg 15:41–55, 1955
5 Martinez-Lage JL: Bony reconstruction in the orbital region. Ann Plast Surg 7:464–479, 1981
6 Edgerton MT, Persing JA, Jane JA: The surgical treatment of fibrous dysplasia, with emphasis on recent contributions from cranio-maxillo-facial surgery. Ann Surg 202:459–479, 1985